 DOI: 10.18231/2455-6750.2016.0006

Parry-Romberg Syndrome: Report of two cases with review of literature

Kundoor V.K. Reddy[1], Modali V. Lakshmi[2], Kotya Naik Maloth[3,*], Kesidi Sunitha[4]

[1]Professor & Head, [2]Post Gradute, [3,4]Senior Lecturer, Dept. of Oral Medicine and Radiology, Mamata Dental College and Hospital, Khammam, Telangana

***Corresponding Author:**
Email: dr.kotyanaik.maloth@gmail.com

Abstract

Parry-Romberg syndrome (PRS) is an uncommon slowly progressing degenerative disease of unknown aetiology, characterized by unilateral atrophic changes on affected part of face involving the skin, fatty tissue, muscles, bone and connective tissue. PRS is a syndrome with diverse complication and associated most common are trigeminal neuralgia, facial paraesthesia, severe headache and epilepsy. Treatment of PRS is a multi-disciplinary approach of various specialities. This article represents such rare cases of PRS in 16 years and 14 years old patients with review of literature.

Keywords: En Coup De Sabre, Hemifacial Atrophy, Parry-Romberg Syndrome.

Introduction

Parry-Romberg syndrome (PRS) also known as progressive hemifacial atrophy is a rare slowly progressing degenerative condition characterized by unilateral atrophy of the face affecting the skin, muscles, connective tissue and bone.(1) In 1825 the Caleb Hillier Parry was the first person to describe it and later Moritz Henirich Romberg described it as a syndrome in 1846 by naming it Prophoneurosis.(2,3) Later in 1871 Eulenberg termed it 'progressive facial hemi atrophy'.(1,2) It is an acquired condition of unknown aetiology, but immune mediated processes is primarily considered. According to Waternberg hyperactivity of sympathetic nervous system, due to disturbed central regulation leads to trophism of underlying fat and subcutaneous tissue. Other possible factors include viral infections, trauma, hormonal disturbances, heredity and autoimmunity.(1,4) Females are more predilection compared to male at a ratio 3:2 and more predilection towards affecting the left side of the face.(1) 50% PRS cases are usually associated with neurological complications such as facial paresthesia, severe headache, trigeminal neuralgia and contralateral epilepsy and 10-35% of cases associated with ocular complication such as Horner's syndrome.(5) Oral manifestations include atrophy of the lip and tongue, decreased height of the body and ramus of mandible, delayed tooth eruption and malformed tooth.(5) Here we report such 2 rare cases of PRS in male patients with oral and neurological problems.

Caser Report 1

A 16 year old male patient visited to the department of oral medicine and radiology with a chief complains of progressive deformity on left side of the face since 4 years. No relevant history regarding medical and trauma was obtained. On physical examination a facial asymmetry with marked hypoplasia on left side of the face with patches of hyperpigmentation on affected skin was noted and in addition ear on left side was slightly smaller than that of right side. (Fig. 1) On intraoral examination, missing 46 and buccally erupted 43 was noted. The occlusion was disturbed and posterior open bite was noted on right side. (Fig. 2) Radio graphically, on panoramic radiography the mandible showed decreased vertical height of ramus and body along with the loss of gonial angle prominence on the left side. (Fig. 3) Posterio-anterior (PA) view and on 3D-CT view showed hypoplasia of mandible, maxilla and mid facial bones on left side. (Fig. 4a and 4b) Based on the history, clinical examination and radiographical findings, the diagnosis of progressive hemifacial atrophy was made.

Fig. 1: Asymmetry on left side of the face

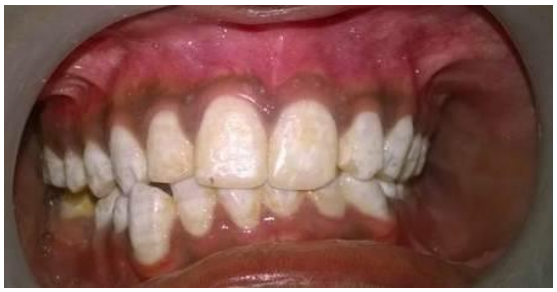

Fig. 2: malocclusion and posterior openbite on right side

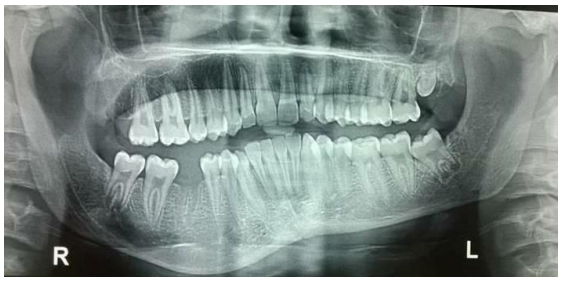

Fig. 3: Orthopantogram revealed decreased vertical height of ramus and body along with the loss of gonial angle prominence on the left side

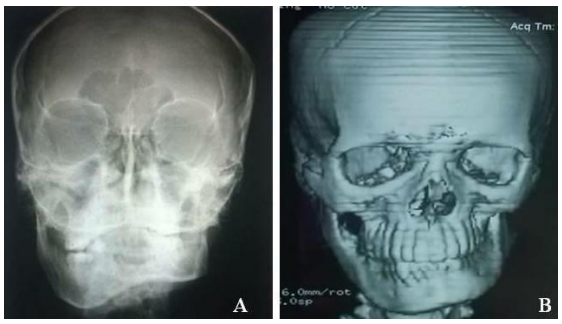

Fig. 4: PA view (a) and 3D CT view (b) revealed hypoplasia of mandible, maxilla and mid facial bones on left side

Caser Report 2

Similar features were noted on right side of the face in a 14 year old boy reported to the department of oral medicine and radiology with a chief complains of progressive deformity of the face since 4 years. Medical history revealed history of epilepsy since childhood and No relevant history regarding trauma. On clinical examination a linear dark scar (coup de sabre) was noted on right side of the fore head (Fig. 5) Radiographically, on 3D CT view showed hypoplasia of mandible, maxilla and mid facial bones on right side. (Fig. 6) Based on the history, clinical and radiographical findings, the diagnosis of progressive hemifacial atrophy was made.

At present both the patient's are being periodically reviewed until atrophic manifestations stops and specific interventions could be accomplished.

Fig. 5: (a) Asymmetry on right side of the face, (b) Big linear scar known as "coup de sabre"

Fig. 5: 3D CT view showed hypoplasia of mandible, maxilla and mid facial bones on right side

Discussion

Parry-Romberg syndrome (PRS) is a rare and slowly progressing degenerative disease, of unknown cause characterized by unilateral atrophy of the facial tissues involving the skin, subcutaneous fatty tissue, muscles, connective tissue and bone.[1] It has a prevalence rate of 1 in 70,000 population and occurs mostly in first and second decades of life with female predominance affecting usually on left side of the face.[3,6] The present cases we report here are male patients and in case 1 affecting on left side where as in case 2 on right side.

The exact aetiology is unknown and remains unclear for this condition, and numerous theories has been proposed by various authors such as autoimmunity, hereditary, trauma, endocrine abnormalities, trigeminal neuritis, sympathetic malfunctions, connective tissue disorders and viral infections.[5,7] Cerebral disturbance on fat metabolism was one of the proposed primary cause for this syndrome.[7] But none of the theories withstands thorough investigations and exact aetiology remains unresolved.

More than an aesthetic concern, this condition brings several functional and psychological problems, when loses symmetrical face and its identity. The extension of atrophy is frequently limited to on one side of the face mostly affecting on left side, and the ipsilateral involvement of body is rare.[1] In the cases presented here; there was involvement of only one side of the face such as case left side and case 2 right side.

On clinical examination, the skin could be dry and hyperpigmented, which is seen in the present cases. In

some patients a demarcation line between normal and abnormal skin, reminding a big linear scar known as "coup de sabre" can be seen,[4,7] as in our case 2. Enopthalmy is a common ocular manifestation, where there is loss of fat around orbit, but the eye functions normally and the ear on the affected side appears smaller due to atrophy than the normal side[3] as was observed in the present cases. Rare representation of neurological problems such as trigeminal neuralgia, epilepsy, facial paraesthesia, and severe headache can be noted.[8] Epilepsy was noted in case 2. Orofacial structures deviation can be noted on the affected side mouth, nose, lips, and tongue. The present cases showed clearly those features of facial asymmetry i.e. except the tongue atrophy, mouth, nose and lips were deviated to affected side.[3,4,8]

Radiographically, on affected side occlusal discrepancy such as unilateral posterior crossbite and posterior open bite, jaw hypoplasia, delayed tooth eruption, incomplete root formations were noted.[9,10] Intraoral soft tissues and masticatory muscles is also involved but the normal function like speech, deglutition are not disturbed.[4,10] The present cases showed posterior open bite and jaw hypoplasia on radiographic findings.

Differential diagnoses included were Bell's palsy, unilateral ankylosis, hemifacial hypertrophy, hemifacial microsomia (1st and 2nd branchial arch syndrome) and its variants such as Goldenhar syndrome, but these are non-progressive diseases. Barraquer Simon syndrome (lipodystrophy), it represents bilaterally with adipose tissue involvement.[3]

Treatment for PRS is a multidisciplinary approach by physicians, surgeons, dentists, psychologists and phonoaudiologists and main goal of therapy is to restore the contour and symmetry of the face. Reposition of the adipose tissue and bone that was lost because of atrophy is the mainstay of the treatment.[3,11] The treatment is usually done are: microvascular surgery, orthognathic surgeries, fat cell injections (lipoinjections), autogenously cartilage, muscle, skin and fat grafts, silicon injections and prostheses, bovine collagen, inorganic implants and recently cell fat mixed with platelet gel are some alternatives to aesthetic correction of the atrophy.[11,12]

Conclusion

The reported cases document the classical features of this rare syndrome, contributing towards the understanding of this rare entity in the management. But till now there was no specific standard guidelines to be followed for treating this rare condition. So a valid treatment protocol needs to be developed in order to establish the best treatment approach with good prognosis.

References

1. Khan M, Khan M, Negi R, Gupta N. Parry Romberg Syndrome with localized scleroderma: a case report. J Clin Exp Dent. 2014;6(3):e313-6.
2. Goldhammer Y, Kronenberg J, Tadmor R, Braham J, Leventon G: Progressive hemifacial atrophy (Parry-Romberg's disease), principally involving bone.J Laryngol Otol 1981,95643.
3. Deshingkar SA, Barpande SR, Bhavthankar JD, Humbe JG. Progressive hemifacial atrophy (Parry Romberg Syndrome). Contemp Clin Dent 2012;3:S78-81.
4. Mazzeo N, Fisher JG, Mayer MH, Mathieu GP, Mcade FGG. Progressive hemifacial atrophy (Parry Romberg Syndrome). Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1995;79:30-5.
5. Whyman RA, Doyle TCA, Harding WJ, Ferguson MM. An unusual case of hemifacial atrophy. Oral Surg Oral Med Oral Pathol 1992;73:564-9.
6. Stone J. Parry Romberg syndrome. Practical Neurology. 2006;6:185-88.
7. da Pinheiro-Silva TP, da Camarinha-Silva C, da Limeira-Silveira CS, Ereno-Botelho PC, Rodrigues-Pinheiro MG, Viana-Pinheiro JJ. Progressive hemifacial atrophy: case report. Med Oral Patol Oral Cir Bucal 2006;11(2):E112-4.
8. Gorlin RJ, Pinborg JJ. Syndromes of the head and neck.McGraw Hill, New York, 1946;pp475-7.
9. Patel H, Thakkar C, Patel K. Parry-Romberg Syndrome: A Rare Entity. J Maxillofac Oral Surg. 2010;9(3):247-50.
10. Foster TD. The effects of hemifacial atrophy of dental growth.Br Dent J 1979;146(5):148-50.
11. Roddi R, Riggio E, Gilbert PM, Houvius SER, Vaandrager JM, vander Meulen JCH. Clinical evaluation of techiniques used in the surgical treatment of progressive hemifacial atrophy. J Craniomaxilofac Surg 1994;22:23-32.
12. Inigo F, Jimenez-Murat Y, Arroyo O, Ferenandez M, Ysunza A. Restoration of facial contour in Romberg's disease and hemifacial microsomia: experience with 118 cases. Microsurgery 2000;20(4):167-72.